and brain stem autonomic functions and a normal sleep–wake cycle, but no signs of awareness. Magnetic resonance imaging of the brain showed diffuse symmetric leucoencephalopathy and brain atrophy. Positron emission tomography showed an overall reduction in resting brain metabolism (especially marked in the bilateral frontal, parietal and cingulate cortex, as well as in the precuneus). The functional magnetic resonance imaging (fMRI) study was carried out in January 2004; the patient's legal guardian provided informed consent. The patient was still in a PVS when he died in 2005.

Event-related fMRI was used to explore brain activity during hearing a phrase (eg, ''Martin, hello Martin'') containing one's own or another first name. Stimuli (normalised to equal sound level) were recorded from 5 men and 5 women, none of which were familiar to the patient or knew his real first name. Two fMRI runs were carried out, both containing 30 own and 30 other name stimuli, and 30 silent null events (duration = 2200 ms; ISI = 1800 ms). During each run, 180 functional images (matrix 64×64, TR 2200 ms, TE 40 ms, FA 86°, 21 six-mm thick slices) were acquired with a Philips 1.5-T MR-Scanner (Philips Medical Systems, Best, The Netherlands). The patient was awake with open eyes during the study.

Statistical parametric mapping software (SPM2; http://www.fil.ion.ucl.ac.uk/spm) was used for data preprocessing (movement and slice-timing correction, linear normalisation to MNI space and 8 mm full-width at half-maximum smoothing) and analysis.

A t test was used to identify regions with higher activity for one's own name than for another name. The same contrast was examined in three male control subjects (aged 32, 42 and 45 years) with a conjunction analysis. Although the control group was not perfectly matched with respect to age, this should not constitute a major problem, as it was mainly included to see whether MPFC activation in response to their own name could be detected with this paradigm. A small volume correction (svc) was used for the MPFC (26 mm diameter sphere, based on previous studies, centred at $x = 2$, $y = 52$, $z = 14$) to correct for multiple comparisons.

The patient showed higher brain activity during hearing his own name than another name in the bilateral MPFC ($x = -6$, $y = 57$, $z = 12$, $p = 0.024$, svc; fig 1A). The same effect was also observed in the left temporoparietal and superior frontal cortex, although only at an uncorrected significance level of $p<0.005$ (fig 1C). We were concerned about normalising the patient's brain owing to the widespread atrophy. However, we used only linear normalisation and manually checked that the results of the normalisation were correct. Additionally, MPFC activation was also found in an analysis using un-normalised data.

The controls showed reliably higher activity for their own names in a comparable MPFC region, restricted to the left hemisphere ($x = -6$, $y = 54$, $z = 22$, $p = 0.044$, svc; fig 1B). At an uncorrected $p<0.005$, this effect was also found in bilateral inferior temporal regions, the right superior parietal and left orbital frontal cortex (fig 1D).

In summary, the patient showed differential cortical processing of his own name compared with another name in the MPFC. A comparable region showed the same effect in three controls. Visual inspection suggests that the MPFC activation was bilateral in the patient, but restricted to the left hemisphere in the controls, although the meaning of this difference remains unclear. At an uncorrected threshold, we identified regions that were activated only in the controls or in the patient. As the small sample size precluded a direct statistical comparison, no conclusions can be drawn on differences in brain activity during this paradigm between this patient with PVS and healthy subjects.

Selective cortical processing of one's own name requires the ability to perceive and access the meaning of words. Clearly, this result shows that residual language recognition can be observed in the PVS, extending previous findings.[1] We do not claim that this result indicates complete awareness, as implicit perception can occur in the absence of conscious perception.

A limitation of this study is that recognising one's own name (an extremely salient stimulus) may be one of the most basic forms of language. This specific response could therefore reflect the existence of an isolated module, which may be compatible with absence of awareness, as connections between distant brain areas, which are linked to consciousness, are disrupted in the PVS.[3]

Furthermore, care should be taken in generalising the results of a single case study, but other fMRI results also indicate that residual language processing can be preserved in the PVS.[4] One could argue that this patient was not in a PVS, but in a minimally conscious state. A selective electroencephalogram response to one's own name in the minimally conscious state was recently reported.[5] However, our patient fulfilled all diagnostic criteria for the PVS, and positron emission tomography showed a compatible metabolic pattern.[3]

Nevertheless, this result may imply that the boundary between different disorders of consciousness is not always clearly delineated. This study shows that fMRI can provide valuable information on residual cognitive processes in the PVS. Information on whether a patient in a PVS can perceive his own name and probably other personally relevant stimuli may also be important for relatives and for planning care and rehabilitative attempts.

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Intracranial hypertension causing polyradiculopathy and late or absent F-waves

Intracranial hypertension is well known to cause cranial neuropathies. The syndrome of intracranial hypertension causing limb weakness and paraesthesias is not, however, well described.[1–4] An illustrative case of intracranial hypertension causing polyradiculopathy with late or absent F-waves is presented and compared with cases previously reported in the literature.

Case report

A 20-year-old previously healthy woman presented with 1 month of intermittent nausea, vomiting, headaches and blurred vision, 1–2 weeks of weakness and paraesthesia in the arms and legs, and pain in the right side of the neck. She was taking no drugs.

Neurological examination showed swollen optic discs. She had bilateral abducens palsies. Shoulder abduction, finger abduction, finger extension and hip flexion were weak bilaterally (Medical Research Council grade 4/5). Muscle stretch reflexes were diffusely absent. Sensation was intact to all modalities. Her gait was slightly unsteady.

Computed tomogram, magnetic resonance angiogram, magnetic resonance venogram and magnetic resonance image of the brain with contrast, and magnetic resonance image of the cervical and lumbar spine with contrast were normal. The following laboratory studies were unremarkable: pregnancy test, electrolytes, complete blood count, liver enzymes, urine analysis, prothrombin time or partial thromboplastin time, antinuclear antibodes and HIV. Erythrocyte sedimentation rate was increased at 60. Lumbar puncture showed an opening pressure of 550 mm H_2O. Analysis of the cerebrospinal fluid (CSF) showed one white cell and one red blood cell, glucose concentration of 78 mg/dl, protein concentration of 26 mg/dl and Gram stain with rare white cells and no organisms.

Visual acuity was OD (L. oculus dexter) 20/50 and OS (L. oculus sinister) 20/40-2 with pinhole. Visual field examination showed mild concentric contraction of isopters for either eye and enlargement of the blind spots.

Nerve conduction studies (NCS) were normal except for absent F-waves in the median, peroneal and tibial nerves and a prolonged minimum F-wave latency in the ulnar nerve (76.5 ms). The right median, ulnar and sural sensory conductions were normal. Electromyography of the right abductor pollicis brevis, first dorsal interosseous, tibialis anterior, gastrocnemius and iliopsoas showed no abnormal spontaneous

Table 1 Cases (n=13) meeting the authors' four criteria

Patient no (ref)	Age/sex	Clinical presentation (in addition to papilloedema)	LP opening pressure (mm H_2O)	LP protein (mg/dl)	Imaging	EDX	Treatment	Outcome
1 (Kincaid 2006)	20/F	HA, CN 6 palsy, paraesthesias, radicular pain, limb weakness and areflexia	1–550	1–26	MRI/MRA/MRV brain—normal	F-waves absent (M, P, T) or delayed (U)	1—Steroids, IVIG	After EVD: radicular pain relief, return of reflexes and strength
			2–550	2–21			2—Diamox, EVD 3—VPS	
2 (Moosa 2004)	24/F	Vision loss, ophthalmoplegia, limb weakness, areflexia, Kerning's sign	>400	40	Digital subtraction angiography —sinus venous thrombosis	Normal, including F-waves	LPS	Improved over 2 weeks
3 (Moosa 2004)	33/F	Neck/back pain, vision loss, NR pupils, ophthalmoplegia, CN 7 palsy, limb weakness, areflexia	>850	44	MRI/MRV Sinus venous thrombosis with haemorrhagic infarct	F-waves absent in all extremities (after LPS: return of F-waves)	LPS	Recovery of CN function and partial return of strength
4 (Kharbanda 2002)	35/F	CN 6 and 7 palsy, limb weakness and areflexia	420	20	CT and MRI brain—normal	F-waves absent	Steroids, diuretics	Gradual recovery but permanent vision loss
5 (Obeid 2000)	24/F	Neck/shoulder pain, CN 6 and 7 palsy, limb weakness and areflexia	420	40	MRI brain and cervical spine—normal	F-waves prolonged (M, U)	1—Steroids 2—IVIG 3—Repeat LPs, diuretics 4—LPS	Improvement with LPS
6 (Obeid 2000)	28/F	L shoulder pain, CN 6 and 7 palsy, limb weakness and areflexia	1–400	1—Normal	MRI/MRA brain—sinus venous thrombosis	F-waves prolonged, decreased amplitude CMAP and SNAP	1—Steroids, diuretics, anticoagulation	Improvement with LPS
			2–570	2–73			2—IVIG 3—LPS	
7 (Weiss 1991)	22/F	Radiating neck pain, paraesthesias, CN 6 and 7 palsy, limb weakness, areflexia	1–600	1–14	CT brain—normal	F-waves absent or prolonged	Repeat LPS and diuretics	Return of strength, but persistent areflexia and papilloedema with changes in visual field
			2–110	2–57				
			3–350	3–62				
8 (Kidron 1989)	18/F	Generalised seizure, bilateral vision loss, ophthalmoplegia, neck stiffness, areflexia	1–600	1–20	CT and MRI brain, cerebral angiogram—normal	ND	1—Steroids, diuretics, EVD	Improvement with EVD; when EVD d/c'd facial diplegia developed; definitive tx with LPS
			2–330	2—Normal			2—LPS	
9 (Kidron 1989)	26/F	Blindness, bilateral opthalmoparesis, bilateral LE areflexia	>600	60	CT brain, cerebral angiogram—normal	ND	1—Steroids, diuretics	Further CN deficits after 3 days of conservative management
							2—EVD	Clinical improvement after EVD
10 (Macaya 1988)	5/F	Neck/back pain with radiation to L arm, hyporeflexia, CN 6 and 7 palsy	1–480	1–16	CT brain—normal	Impersistent F-waves (M, P, T) normalised after repeat LPs	Repeat LPS	Improved radicular pain after LP, resolution of papilloedema
			2–510	2–23				
11 (Murray 1986)	4/F	L neck/shoulder pain, L arm areflexia	1–550	1–16	CT brain, cervical myelogram, bone scan—normal	ND	1—Repeated LPS	Pain resolved after repeated LP, asymptomatic with normal examination at 5 months
			2–550 3–350	2 and 3—Normal			2—Steroids, diuretics	

Table 1 Continued

Patient no (ref)	Age/sex	Clinical presentation (in addition to papilloedema)	LP opening pressure (mm H_2O)	LP protein (mg/dl)	Imaging	EDX	Treatment	Outcome
12 (Bortoluzzi 1982)	34/F	Back/leg pain, numbness T6–T12, LE weakness, global hyporeflexia	350–450 (Multiple)	Normal	CT brain, spine—normal, cisternography—enlarged SA space below L2	ND	1—Steroids, diuretics 2—LPS Steroids	Improved pain after LPS, normal examination at 7 months
13 (Sullivan 1977)	24/F	CN 6 and 7 palsies, limb weakness, unilateral hyporeflexia	1–310	1–15	CT brain, cerebral angiogram, normal	F-waves—ND		Gradual resolution of papilloedema and facial palsy
			2–245	2–23				

CMAP, compound muscle action potential; CN, cranial nerve; CT, computed tomography; d/c'd, discontinued; EDX, electrodiagnostic testing; EVD, external ventricular drain; HA, headache; IVIG, intravenous immunoglobulin; L, left; LE, lower extremity; LP, lumbar puncture; LPS, lumboperitoneal shunt; mm H_2O, millimetres water; M, median nerve; MRA, magnetic resonance angiogram; MRI, magnetic resonance imaging; MRV, magnetic resonance venogram; ND, not done; NR, non-reactive; P, peroneal nerve; R, right; SA, subarachnoid; SNAP, sensory nerve action potential; T, tibial nerve; tx, treatment; U, ulnar nerve; VPS, ventriculoperitoneal shunt.

activity and normal motor unit action potentials. A neurogenic recruitment pattern was present in the abductor pollicis brevis muscle.

The patient's vision and weakness worsened despite treatment with intravenous immunoglobulin for the presumed diagnosis of Guillain–Barré syndrome (GBS). Repeat ophthalmological examination showed loss of nasal field in the left eye. Repeat lumbar puncture (5 weeks after the onset of symptoms) showed an opening pressure of >550 mm H_2O with a protein concentration of 21 mg/dl in the CSF. NCS continued to show prolonged or absent F-waves without other demyelinating features. The patient was started on prednisone and acetazolamide and an external ventricular drain was placed. During the first 24 h after external ventricular drain placement, intracranial pressure was greater than 400 mm H_2O. The patient's nausea, headache and neck pain resolved. Visual blurring and distal paraesthesias improved over several days. Intracranial pressure dropped to <200 mm H_2O, and a ventriculoperitoneal shunt was placed. Over the next 24 h, muscle stretch reflexes returned in the lower extremities and hip flexor strength improved. NCS showed return of normal F-wave responses. Prednisone and diuretic treatment were discontinued.

A follow-up clinic visit 2 weeks later found that the patient had improved. Her examination showed only a mild right abducens palsy and minimal hand weakness. She continued to have bilateral papilloedema. Telephone follow-up 1 and 4 months later showed that she was asymptomatic.

Literature review

The authors reviewed the literature (Pub Med) for case reports meeting the following criteria: (1) papilloedema, (2) increased opening pressure on lumbar puncture (>250 mm H_2O), (3) normal (<45 mg/dl) or slightly increased (45–75 mg/dl) protein concentration in the CSF and (4) areflexia or hyporeflexia.

Including our case, 13 cases met these criteria, 10 with benign intracranial hypertension and 3 with cerebral venous sinus thrombosis (table 1).[1–9] A total of nine patients were reported to have limb weakness, two with tetraplegia. Radicular pain or paraesthesias were described in nine patients. Cases of abducens palsy (n = 7), bilateral global ophthalmoplegia (n = 4) and facial weakness (n = 7) were reported. Headache and visual loss were common, absent in only three patients.

Placement of a CSF shunt led to clinical improvement characterised by return of strength, reflexes, F-waves or elimination of radicular pain in all eight patients.

In the five patients who did not undergo continuous CSF drainage, three showed clinical improvement (patients 10, 11 and 13), but two showed persistent visual loss (patients 4 and 7).

Discussion

We propose that intracranial hypertension can cause a syndrome of weakness and areflexia in addition to the more classic signs of intracranial hypertension. Our patient and seven other patients reported absent or prolonged F-waves on NCS in the absence of other demyelinating features. This finding is often considered to be an early electrodiagnostic finding in GBS. Follow-up NCS were carried out in our patient and in two patients (patients 3 and 10), none of whom developed other electrophysiological evidence of demyelination and all showed return of normal F-wave latencies after definitive treatment of intracranial hypertension. The return of normal F-waves after CSF shunting or repeated lumbar puncture is supportive of a cause and effect relationship between intracranial hypertension and polyradiculopathy. On follow-up, protein concentrations in the CSF continued to be normal or slightly increased in all cases reporting follow-up CSF results (mean 39.2 mg/dl, range 21–73), again arguing against the theory that these cases represent GBS with papilloedema.

Patient 12 initially underwent back surgery for persistent radicular pain. During surgery, the authors noted the presence of ''enlarged'' nerve roots and lumbar dural sac. A cisternogram also documented enlargement of the subarachnoid spaces below L-2 and of the thoracic root pouches bilaterally.[7] The mechanism by which intracranial hypertension causes weakness and areflexia is unknown, but this finding would argue in favour of the theory that increased pressure in the subarachnoid spaces, with subsequent compression of the nerve roots, is the cause of the polyradiculopathy.

In conclusion, we propose that in particularly severe cases of intracranial hypertension, an under-recognised syndrome of polyradiculopathy with absent or prolonged F-waves may result. CSF shunting may lead to a better outcome than medical management in these patients, particularly with regard to preservation of vision. Therefore, timely recognition of this syndrome is necessary.

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